Unforeseen complications of Sickle cell disease & its challenging management.

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Case Discussion

Background

- Patient was diagnosed with HbSS variant in childhood. Experienced significant recurrent episodes of sickle cell crisis & its complication; end organ damage (CKD stage 3a/A2), leg ulcers.
- Baseline Hemoglobin 5-6g/dL. On hydroxyurea 500/1000mg alternating daily. Blood transfusion was avoided due to alloimmunization, transfused only if hemodynamically unstable. Started on Aranesp (Darbepoetin Alfa) 0.4mcg/kg once every 2-weeks. Hb F 1.3%, ferritin >1500g/dL, Hb 4.1g/dL, MCV 108.2fL, retic % 16.3, LDH 685 IU/L, EPO 143 mIU/ml.
- For pain management on oxycodone due to CKD stage 3a/A2.
- While sickle cell disease/Acute chest syndrome and pericarditis/cardiac tamponade are not associated in the current medical literature. The pathophysiology requires further investigation.
- Idiopathic pericarditis is uncommon in SCD. NSAIDs are the 1st line treatment. Pericardial effusion impending tamponade requires pericardial drain & if persistent pericardial window should be considered.
- Oxidative stress plays an important role in complication of SCD, hence role of arginine, NSAIDs, steroids, allopurinol have been suggested. Butyrate (fatty acid) has shown increase in HbF in some studies. Inhibitors of acetone deacetylase (HDAC) have shown to decrease vascular complications of SCD.
- Red cell exchange transfusion is an under utilized but highly effective treatment for both chronic & acute complications of SCD by decreasing blood viscosity & increasing oxygen carrying capacity of blood. Whereas simple transfusion only improves oxygenation.
- Iron neutrality is maintained in red cell exchange transfusion as the removed erythrocytes are replaced with non-immunogenic units. Allopurinol was added to the exchange transfusion.
- A complete effective communication during transfer of care is vital in effective management of patient with multiple rare complications.

Discussion

- Arthritis associated with SCD are usually symmetrical 60%, polyarticular 80%, involving large joints of lower extremity. Periartricular erosions, bony infarcts, synovitis can be seen on X-ray. 1,2
- Conservative management of arthritis involves analgesics, hydration, physical therapy. Followed by core decompression & eventually arthroplasty. In case of septic arthritis immediate joint debridement & irrigation should be performed. OM requires IV antibiotics. 3

References